JOURNAL OF MEDICAL CASE REPORTS

CASE REPORT Open Access

Novel two-stage surgical treatment for Cantrell syndrome complicated by severe pulmonary hypertension: a case report

Xiaochen Wu[1], Jinbao Zhang[1*], Hui Ouyang[1], Qin Yue[1] and Heng Zhao[2]

Abstract

Introduction: Cantrell syndrome is a rare syndrome of congenital defects, which can be complicated by severe pulmonary hypertension and left ventricular diverticulum; it has proved difficult to treat in clinical practice.

Case presentation: A 6-month-old Han Chinese baby girl weighing 3.5kg was diagnosed, using ultrasonography and radiography, as having Cantrell syndrome complicated by severe pulmonary hypertension. For safety, we divided management into two stages. For the first stage, we dealt with the left ventricular diverticulum and pulmonary hypertension. Three months later, we performed diorthosis for an intracardiac malformation.

Conclusions: Cantrell syndrome with pulmonary hypertension may respond well to this novel two-stage operation, which needs more verification via clinical practice.

Keywords: Cantrell syndrome, Left ventricular diverticulum, Pulmonary hypertension

Introduction

Cantrell syndrome is a rare syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium, and heart. The spectrum of anomalies varies widely, ranging from incomplete to severe expression with involvement of other organ systems [1,2]. Cantrell syndrome with severe pulmonary hypertension is quite rare, and a case complicated by left ventricular diverticulum is extremely rare. The cardiac anomaly and severe pulmonary hypertension in Cantrell syndrome increases operative mortality. It is clinically difficult to safely and completely cure the syndrome [3]. We report a rare case of Cantrell syndrome complicated by severe pulmonary hypertension and left ventricular diverticulum in a patient who survived two-stage operative procedures.

Case presentation

A 6-month-old Han Chinese baby girl weighing 3.5kg with a large (5 × 5cm) intact omphalocele over her anterior abdominal wall (Figure 1) was brought to our hospital by her parents who denied heart disease history. Her parents were scared because of her large intact omphalocele after full-term normal delivery and worried that she would develop heart disease. The final diagnosis was made by radiography and ultrasound detection. Because of dextrocardia, the patient's cardiac apex was located in the fourth intercostal space on her right side. No murmurs indicating severe pulmonary hypertension were auscultated. The rest of her physical examination was normal. Radiography showed dextrocardia and a huge omphalocele (Figure 2, red arrow). Her sternum was short, with defective formation of its lower third. Echocardiography (Figure 3) also revealed dextrocardia, an ostium secundum atrial septal defect (ASD) measuring 0.4cm with bidirectional shunt and a 1.2cm ventricular septal defect (VSD) with main right-to-left shunt. Patent ductus arteriosus and left ventricular diverticulum were discovered. The extent of her left ventricular diverticulum was to the omphalocele and ended with a cecum. The final diagnosis of Cantrell syndrome that was suspected by physical examination was confirmed by ultrasonography and radiography. She underwent two-stage surgery after 1 month of medication with prostaglandin E. Her legal guardian gave consent for each of the treatments including surgery and drug management for the duration of her hospital stay.

* Correspondence: nowwilson@yeah.net
[1]Department of Cardiovascular, Chengdu Millitary General Hospital, 270th Rongdu Road, Chengdu City 610083, China
Full list of author information is available at the end of the article

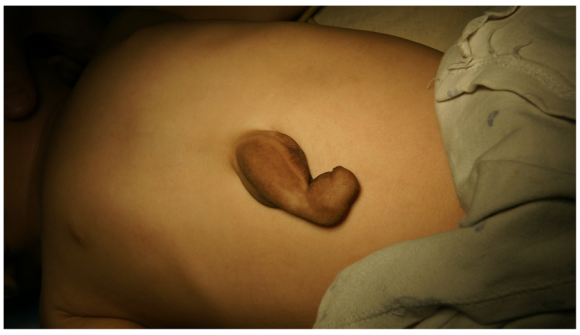

Figure 1 Large intact omphalocele on the anterior abdominal wall. The omphalocele measured 5 × 5cm.

The first stage of the treatment was aimed at improving cardiac function and controlling her pulmonary hypertension before subjecting her to intracardiac repair. Adequate pulmonary flow control is a useful and effective option for any patient with pulmonary hypertension. In addition, we aimed to help her gain weight by improving her nutrition, which would reduce the operative risk when we finally undertook the repair. When she seemed to be in better shape, we performed bilateral pulmonary artery (PA) banding through a median sternotomy. We instituted banding of 6mm circumference for bilateral PAs using our handmade banding tape in which an expanded polytetrafluoroethylene sheet of 0.4mm thickness was trimmed to 3.0mm width. The degree of banding may be determined by changes in oxygen desaturation. When the fraction of inspired oxygen (FiO_2) was 1.0, oxygen desaturation was less than 60%. By loosening the right circumference from 12mm to 14mm, the saturated oxygen ($SatO_2$) level increased to 81% at FiO_2 80%. Finally, the circumferences of the right and left PA banding were fixed at 8mm and 6mm, respectively. We then ligated her left ventricular diverticulum and patched defects of her diaphragm and exomphalocele. Extubation was performed 3 hours after the operation. Inotropic support was completed 20 hours postoperatively.

Three months after completion of the first-stage surgery stage 1, with uninterrupted prostaglandin E administration throughout, her $SatO_2$ stabilized at 85% in room air. On that basis, we decided to perform the second operation through a median sternotomy. After extracorporeal circulation was established, the ductus arteriosus was sutured through a pulmonary trunk incision with 6–0 Prolene. Then, her ASD and VSD were patched through a right atrial incision. Both ASD and VSD patching were done with bovine pericardium sheet and 6–0 Prolene in running sutures. Her postoperative clinical course was not smooth.

After 14 days of continuous intubation, she was extubated and was discharged from the hospital. At 6 months postoperatively, however, she died from respiratory failure caused by a severe pulmonary infection.

Discussion

In 1958, Cantrell *et al.* [3] described a rare syndrome that is characterized by the following anomalies: a supraumbilical abdominal wall defect (omphalocele or exomphalocele), lower sternum, a defect of the anterior diaphragm, dextrocardia, and congenital intracardiac defects, among others. The prognosis for patients with Cantrell syndrome is dependent on the severity of the omphalocele, cardiac

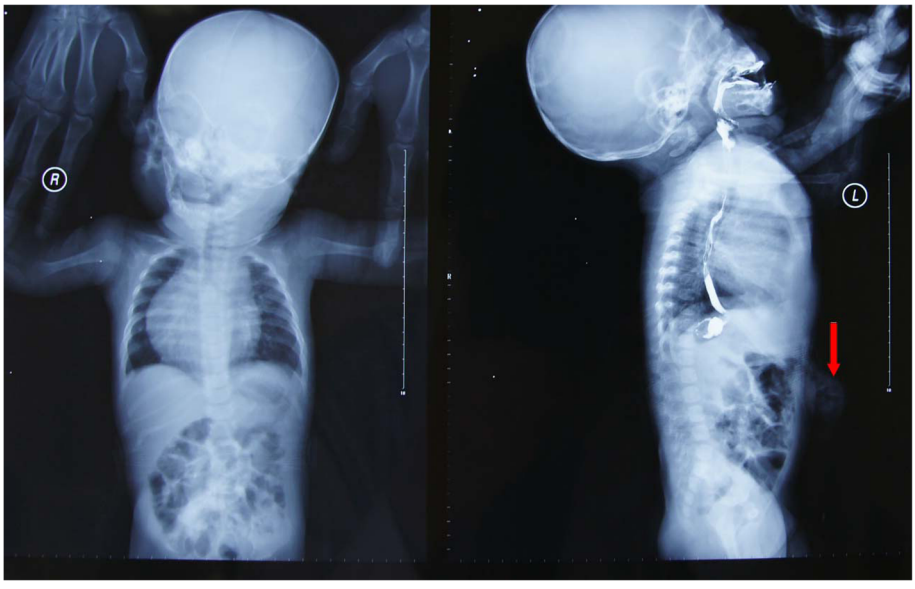

Figure 2 Radiography showing dextrocardia and a large omphalocele (arrow). R, Right; L, Left.

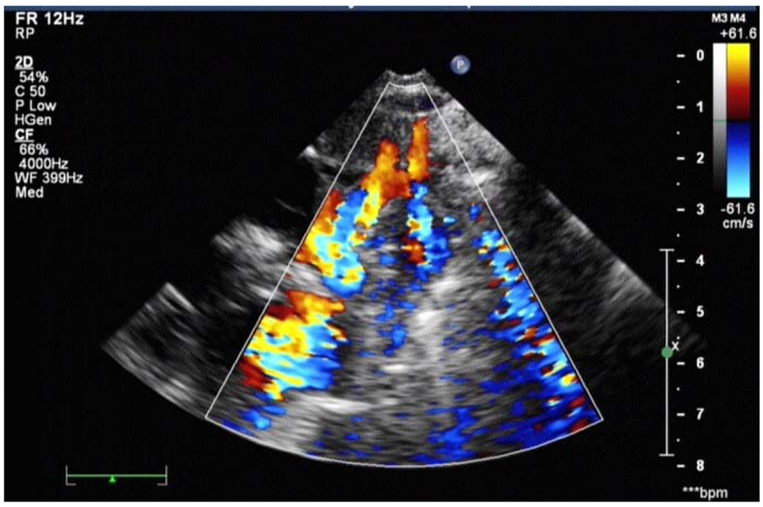

Figure 3 Echocardiography revealing abnormalities. Abnormalities include dextrocardia, an ostium secundum atrial septal defect with bidirectional shunt, and a ventricular septal defect with main right-to-left shunt.

anomalies, and dextrocardia [4]. A detailed review of the literature indicates that the syndrome occurs in varying degrees of severity, from incomplete to severe expression with involvement of other organ systems [2]. Intracardiac defects, such as VSD, ASD, tetralogy of Fallot, and ventricular diverticulum, are common in Cantrell syndrome, whereas pulmonary hypertension is rare [5]. Based on the previously reported severity grades, our patient fell into the high-risk group.

For our case, because of severe pulmonary hypertension, we preferred a two-stage operation. Controlling unstable pulmonary flow by main PA banding has been reported for truncus arteriosus [6], although it would not be indicated for all anatomical types. Bilateral PA banding can be used for patients with bilateral PA. During the first stage of treatment, we performed bilateral PA banding, ligated the left ventricular diverticulum, and patched defects of the diaphragm and exomphalocele. Everything went well during this stage. Three months later, for the second stage we sutured the ductus arteriosus and patched the ASD and VSD defects. She was extubated 14 days postoperatively. She survived this stage but died 6 months later of respiratory failure due to a pulmonary infection. For this very rare case, we found that a two-stage surgical program was effective. It is our goal in clinical research to increase the survival rate.

Conclusions

Cantrell syndrome, which is diagnosed by ultrasonography and radiography, including computed tomography [7], is a rare syndrome of congenital defects. There is no ideal treatment regimen for it. In this case, a two-stage surgical plan was approved. It was a novel and effective treatment introduced to provide patients with a longer survival period and a higher survival rate.

Consent

Written informed consent was obtained from the patient's legal guardian for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Abbreviations

ASD: Atrial septal defect; FiO_2: Fraction of inspired oxygen; PA: Pulmonary artery; $SatO_2$: Saturated oxygen; VSD: Ventricular septal defect.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

XW, JZ, HO, and QY participated in the design of the study and carried out the operation. XW took care of the patient during the surgical part of the treatment and drafted the manuscript. HZ performed the ultrasonography scanning. All authors read and approved the final manuscript.

Authors' information

Xiaochen Wu, first author, MD, Cardiovascular Surgery; Zhang Jinbao, corresponding author, PhD, Cardiovascular Surgery and Director of the Cardiovascular Department; Ouyang Hui, PhD, Cardiovascular Surgery; Heng Zhao, MD, Ultrasonography; Qin Yue was in charge of cardiopulmonary bypass during the cardiac operation.

Acknowledgment

This work was supported by the Chendu Military Science Fund (C12039). The authors thank Prof. Zhang Guizhen for critical review and assistance with manuscript preparation.

Author details

[1]Department of Cardiovascular, Chengdu Millitary General Hospital, 270th Rongdu Road, Chengdu City 610083, China. [2]Department of Ultrasound, Chengdu Millitary General Hospital, 270th Rongdu Road, Chengdu City 610083, China.

Received: 24 June 2013 Accepted: 27 January 2014
Published: 26 March 2014

References

1. Kinoshita M, Park S, Shiraishi T, Ueno S: **Thoracoabdominoplasty with umbilicoplasty for Cantrell's syndrome.** *J Plast Surg Hand Surg* 2012, **46**(5):367–370.
2. Vanamo K, Sairanen H, Louhimo I: **The spectrum of Cantrell's syndrome.** *Pediatr Surg Int* 1991, **6**:429–433.
3. Cantrell JR, Haller JA, Ravitch MM: **A syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium and heart.** *Surg Gynecol Obstet* 1958, **107**:602–614.
4. Quandt D, Dave H, Valsangiacomo BE: **Heart with a trunk: form fruste of Cantrell's syndrome.** *Eur Heart J* 2011, **32**(1):123.
5. Manieri S, Adurno G, Iorio F, Tomasco B, Vairo U: **Cantrell's syndrome with left ventricular diverticulum: a case report.** *Minerva Pediatrics* 2013, **65**(1):93–96.
6. McFaul RC, Mair DD, Feldt RH, Ritter DG, McGoon DC: **Truncus arteriosus and previous pulmonary arterial banding: clinical and hemodynamic assessment.** *Am J Cardiol* 1976, **38**:626–632.
7. Angoulvant D, Sanchez I, Boussel L: **Late diagnosis of incomplete Cantrell's syndrome on CT scan.** *Arch Cardiovasc Dis* 2011, **104**(3):208–210.

doi:10.1186/1752-1947-8-107
Cite this article as: Wu *et al.*: Novel two-stage surgical treatment for Cantrell syndrome complicated by severe pulmonary hypertension: a case report. *Journal of Medical Case Reports* 2014 **8**:107.